Modern Concepts of Cardiovascular Disease

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# ENLARGEMENT OF THE HEART IN INFANTS AND YOUNG CHILDREN

The cause of enlargement of the heart in infants and young children is still a fertile field for investigation. Some long-used terms need to be restudied and re-evaluated in the light of the increased knowledge now open to us because of the development of instruments of precision, and the advance of studies in pathology and chemistry. An example of such need is seen in the use of the terms "idiopathic hypertrophy," and "congenital idiopathic hypertrophy." Only in recent years has it been demonstrated that many of these cases were not genuine examples of idiopathic hypertrophy of the heart, since either myocardial disease or other factors were found which could have had a causal relationship to the cardiac enlargement.

In most instances the cause or nature of the enlargement of the heart in infants or young children can be determined if the criteria of diseases, as outlined below, be kept in mind:

### Classification and Differential Diagnosis

**I. Congenital Defects:** (a. Heart, b. Coronary arteries, c. Aorta and pulmonary artery.)

In the first few years of life, cardiac hypertrophy is frequently due to a congenital malformation. In children with congenital heart disease, symptoms are apt to present themselves early, particularly in the group with a venous-arterial shunt. Here cyanosis is an early sign, and murmurs are generally loud and rumbling. Roentgenological examination of the heart may reveal that its outline is bazarre. The electrocardiogram may show evidence that will confirm the diagnosis of congenital heart disease. Congenital anomalies of the coronary arteries, such as malposition or maldevelopment, which cause myocardial degeneration, may be followed by marked enlargement of the heart.

**II. Infections:** (a. Unknown etiology: Rheumatic fever, Fiedler's myocarditis, Periarteritis nodosa, etc.; b. Known etiology: Diptheria, Scarlet fever, other acute bacterial infections; Subacute bacterial endocarditis with valvular defect, Syphilis, etc.)

Enlargement of the heart caused by rheumatic infection may be found early in childhood. It occurs, indeed, earlier and more frequently than is generally appreciated. A familial history of rheumatic fever should arouse suspicion of the presence of this infection in a child with cardiac enlargement. The signs and symptoms are: fleeting joint or muscle pains, spasmodic twitchings (choreiform movements), abdominal manifestations such as cramps or, at times, symptoms simulating intra-abdominal inflammation. Evidence of tonsillitis, pharyngitis, polyarthritis, or myocarditis may be found. There may be continued fever, leucocytosis, abnormal sedimentation rate, and electrocardiographic changes. The murmurs in markedly enlarged hearts are usually loud and rumbling. Roentgenological examination may show typical "mitralization" of the heart, with an enlarged left auricle.

Fiedler's myocarditis is extremely rare in infants and young children, but it can occur and therefore should be kept in mind. Such patients may have low-grade fever and definite electrocardiographic changes. The heart is the seat of an acute myocarditis, and the lesions are characteristic.

The heart may be enlarged when affected by diphtheria, scarlet fever, bacterial endocarditis, syphilis, and other bacterial infections. The problem of differential diagnosis here is a question of the recognition, or the exclusion, of infections of known bacterial origin. The question of other rare forms of myocarditis arises only when the history indicates such a possibility, (Trichinosis, etc.)

**III. Anemias.** (Long standing; a. Primary, b. Secondary.)

Enlargement of the heart in infants and young children can occur in severe anemia. Apical systolic murmurs may be heard. The hearts may return to normal size and the murmur disappear when the hemoglobin returns to normal.

**IV. Syndrome of "Non-suppurative Myocardial Degeneration with Dilatation and Hypertrophy."**

In 1933 we described seven cases of unusual enlargement of the heart in infants and young children. In all these the clinical picture and the pathological changes were similar. Later, further investigation of fresh pathologic material, with chemical examinations in another similar case confirmed the impression that this form of cardiomegaly was different from Von Gierke's Disease and other forms of cardiac enlargement.

In our classification of cardiac enlargement in children we have designated this group as the "Syndrome of non-suppurative myocardial degeneration with dilatation and hypertrophy." Clinically it is characterized by an unusual degree of enlargement of the heart, an afebrile course, abnormal electrocardiographic changes, a tendency to abrupt onset of myocardial failure, and sudden death.

The early symptoms of this disease may be rapid respiration without apparent intrathoracic cause. Dyspnea and cyanosis, however, are usually of sudden onset. The temperature remains normal unless there are complicating independent infections. The heart rate is usually rapid, and the heart sounds of poor quality. At times a soft, blowing, systolic murmur may be heard at the apex. In the terminal stages the liver is enlarged. Rales may be

heard in the chest, and at times edema occurs. The most striking fact about this entire clinical picture is that the infants or children were usually apparently well and then suddenly developed myocardial failure, cried, vomited, or refused food.

Unless the chest be examined carefully, the true nature of this condition may not be appreciated. Even the cardiac enlargement may not be discovered by physical examination, but roentgenograms of the chest usually will reveal a surprising degree of enlargement. This can be so great that the heart may fill almost completely the left side of the chest in its transverse diameter. Although the roentgenographic appearance is strikingly suggestive, it is by no means pathognomonic. A similar shadow may be cast in dilatation and hypertrophy of the heart in cases of valvular disease, congenital anomalies, or marked anemia. However, the enlargement of the left auricle and the rumbling murmurs of mitral stenosis, found in rheumatic cardiovalvular disease, are not found. Two of these patients were examined by means of the electrocardiograph and both showed evidence of myocardial damage, such as low voltage, shallow T waves, or prolongation of the P-R interval.

The pathological lesions were similar in all the cases. The hearts were dilated and hypertrophied and the weights greatly increased. The endocardium of the left auricle and left ventricle was sometimes thickened. At times bland thrombi were found attached to the ventricular endocardium. There were no valvular or congenital defects. On cutting the myocardium there were seen, in some instances, grayish streaks which on microscopic examination proved to be foci of atrophied and degenerated muscle fibres. Occasionally a few lymphocytes were found. There were no suppurative foci. The heart muscle arrangement was in some instances so distorted that it was difficult to recognize. The coronary arteries showed only slight changes.

The etiology of this disease is at present undetermined. The only consistent factor in our cases has been a possible previous infection. In most of the cases the disease has been recognized before the second year of life. It affects both sexes and both the white and colored races. We have observed two similar cases in adults.

At present little can be said concerning treatment. All eight patients died, six within less than a week after admission. However, our observations in one child and in two adults in which symptoms of myocardial failure lasted from ten weeks to three years, give some hope for prognosis and therapy. Prolonged rest, instituted early, and the administration of proper doses of digitalis may have helped in some of these cases.

V. **Metabolic Disturbances.** (a. Avitaminosis, b. Thyroid deficiency, c. Von Gierke's Disease, etc.)

In the Orient, or in areas where beriberi is prevalent, cardiac enlargement in children is a frequent finding. These patients do not respond to the ordinary treatment for cardiac failure, but do recover when vitamin B is administered.

Von Gierke's Disease is due to a disturbance of dextrose metabolism and is characterized clinically by enlargement of the liver, kidneys or heart. The cause of the enlargement is the accumulation of glycogen in abnormal amounts in the parenchymal cells.

Clinically there are two types; hepatic and cardiac. The clinical manifestations depend upon the extent of the damage to the involved organs. In the cardiac cases there may be a markedly enlarged heart with progressive myocardial failure, manifested by rapid breathing, cyanosis, edema, and generalized weakness. The electrocardiographic findings in the case of Dr. Paul White were normal. A normal electrocardiogram in addition to chemical studies may help in the diagnosis of Von Gierke's Disease in infants and young children. In the later age period, however, the electrocardiogram may show evidence of the enlargement of the heart as well as myocardial damage (Dr. Arthur Master). In the hepatic cases, acetonuria or ketonuria may occur. Adrenalin fails to induce an appreciable increase in blood sugar or blood lactic acid, or to diminish ketonuria. The blood sugar in the fasting states has been found to be low. Chemical studies should be made in all cases of enlargement of the heart to rule out Von Gierke's Disease. The Pathological lesions are characteristic.

VI. **Hypertension.** (a. Greater circulation: Essential; adrenal tumors; secondary to kidney lesions —inflammatory or congenital; b. Lesser circulation: lesions in lung, kyphoscoliosis, etc.)

Enlargement of the heart associated with hypertension, either primary or secondary, has been found in children. Hypertension following rheumatic fever or associated with suprarenal tumors presents a definite clinical picture for differential diagnosis. In cases of spinal deformities, hypertrophy and dilatation of the heart may occur. In these instances the enlargement of the heart is usually right-sided, and is believed to be secondary to hypertension of the lesser circulation.

VII. **Tumors of the Heart.** (a. Primary, b. Secondary.)

VIII. **Unclassified Group.**


### Summary

Only in recent years has it been demonstrated that in many cases what was formerly called "idiopathic hypertrophy" of the heart was in reality associated with congenital malformations, rheumatic fever, glycogen-storage disease, myocardial degeneration and fibrosis, etc. Cases of dilatation and hypertrophy of the heart, associated with myocardial degeneration and fibrosis, constitute the majority of those formerly included under the title of "idiopathic hypertrophy." The term "congenital idiopathic hypertrophy" of the heart is not only undesirable but also confusing.

A differential diagnosis of cardiomegaly in children can be made if one bears in mind the various criteria of diseases as outlined. Such classification and differentiation are not merely of academic interest. It is important to realize that an enlarged heart is usually a diseased heart.

The only hope for the development of effective therapy lies in the early recognition and diagnosis of these cases.



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